Title
A green-brown plaque on the dorsal hand.

Journal
JAAD case reports, 5(1)

ISSN
2352-5126

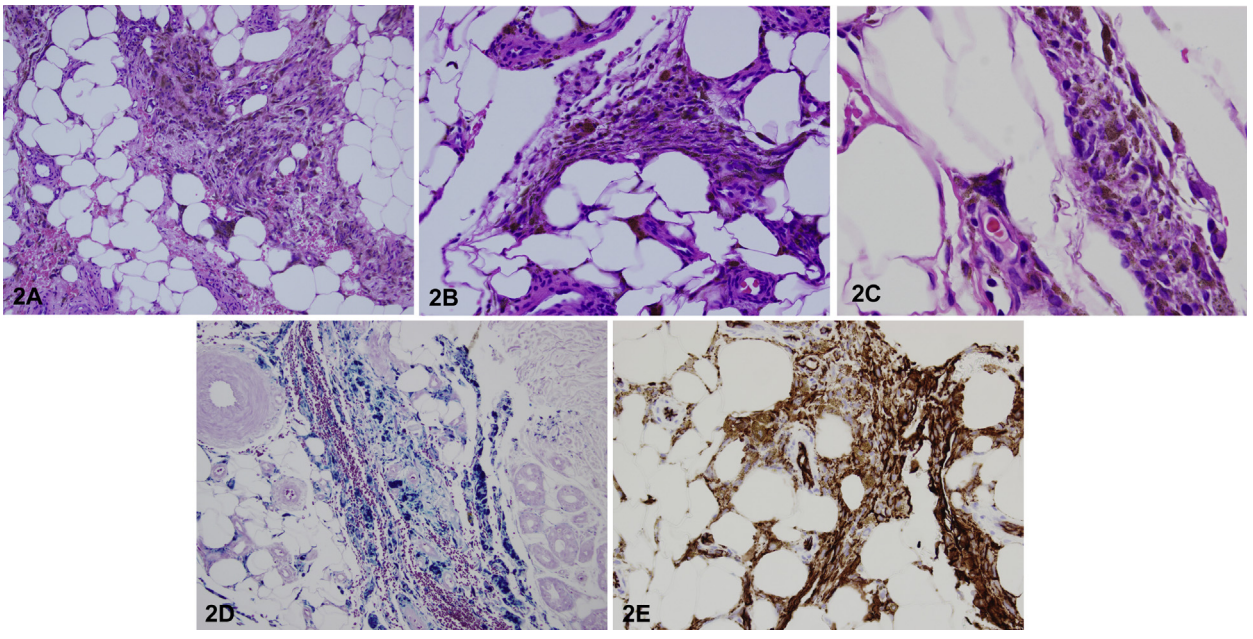
Authors
Hosking, Anna-Marie
Pouldar, Delila
Elsensohn, Ashley
et al.

Publication Date
2019

DOI
10.1016/j.jdcr.2018.09.011

A green-brown plaque on the dorsal hand

Anna-Marie Hosking, BS, Delila Pouldar, MD, Ashley Elsensohn, MD, MPH, Sama Kassira, MD, Janellen Smith, MD, and Sebastien de Feraudy, MD, PhD
Irvine, California



From the Department of Dermatology, University of California, Irvine.
Funding sources: None.
Conflicts of interest: None disclosed.
Correspondence to: Anna-Marie Hosking, BS, Department of Dermatology, University of California, Irvine, 118 Med Surge I, Irvine, CA 92697. E-mail: ahosking@uci.edu.

JAAD Case Reports 2019;5:60-2.
2352-5126

https://doi.org/10.1016/j.jdcr.2018.09.011

A 62-year-old man presented with a 1-year history of a green-colored plaque over his left dorsal hand (Fig 1). The patient denied any history of trauma, and there were no associated symptoms, including pain or pruritus. The discolored area was stable in size, and the patient denied any fever, chills, night sweats, or weight loss. Physical examination of the left dorsal hand found a green-brown plaque measuring 4 cm by 3 cm. A punch biopsy specimen was obtained and staining was performed, including hematoxylin-eosin (Fig 2, *A-C*), iron special stain (Fig 2, *D*) and CD34 (Fig 2, *E*). Definitive surgery was completed after histopathologic review.

Question 1: What is your diagnosis?

A. Spindle cell lipoma

B. Dermatofibrosarcoma protuberans (DFSP)

C. Hemosiderotic fibrolipomatous tumor (HFLT)

D. Spindle cell hemangioma

E. Epithelioid sarcoma

Answers:

A. Spindle cell lipoma — Incorrect. Spindle cell lipoma more commonly occurs on the neck, shoulder, or back. Histopathologically, spindle cell lipoma presents as an adipocytic tumor with variable admixture of spindle cells but, unlike HFLT, has a prominent myxoid stroma and bundles of ropy collagen and lacks abundant hemosiderin.[1]

B. DFSP — Incorrect. DFSP presents as a slow-growing large nodule or plaque, most commonly on the trunk or extremities. On histopathology, DFSP also shows a $CD34^+$ spindle cell proliferation, often arranged as whorls (so-called storiform pattern), but does not exhibit prominent hemosiderin deposition, the prominence of adipocytes, or inflammation.[1]

C. HFLT — Correct. HFLT presents as a slow-growing faintly green plaque on the dorsal aspect of the distal extremities, most commonly involving the ankle or foot. Histopathologically, HFLT is a nonencapsulated fatty lesion composed of short fascicles of $CD34^+$ spindle cells with vesicular, occasionally hyperchromatic, nuclei and indistinct nucleoli, some multinucleated cells, disposed between mature adipocytes within an inflammatory background containing abundant hemosiderin deposition. Numerous thick-walled vessels may also be present.[1,2]

D. Spindle cell hemangioma — Incorrect. A spindle cell hemangioma commonly presents as a subcutaneous mass on the distal extremities and could account for the $CD34^+$ spindle cell proliferation with abundant hemosiderin deposition, but spindle cell hemangioma also presents with large, dilated cavernous spaces. The presence of mature adipocytes admixed with spindle cells does not fit this type of tumor.[1]

E. Epithelioid sarcoma — Incorrect. Although epithelioid sarcoma also presents as $CD34^+$, slow-growing firm subcutaneous nodules that commonly involve the distal extremities (hands), prominent hemosiderin deposition, and mature adipocytes are not present.[1]

Question 2: Which of the following immunohistochemistry staining profiles is characteristic of this tumor?

A. $CD34^+$, $desmin^-$, smooth muscle actin $(SMA)^+$, $keratin^-$, $S100^+$

B. $CD34^+$, $desmin^-$, SMA^+, $keratin^-$, $S100^-$

C. $CD34^+$, $desmin^-$, SMA^-, $keratin^-$, $S100^-$

D. $CD34^-$, $desmin^-$, SMA^-, $keratin^+$, $S100^+$

E. $CD34^-$, $desmin^+$, SMA^-, $keratin^+$, $S100^+$

Answers:

A. $CD34^+$, $desmin^-$, SMA^+, $keratin^-$, $S100^+$ — Incorrect. The S100 stain is used to differentiate melanocytic from nonmelanocytic lesions. Most melanomas are $S100^+$, whereas, HFLT is $S100^-$.[3]

B. $CD34^+$, $desmin^-$ SMA^+, $keratin^-$, $S100^-$ — Incorrect. SMA is a marker for myofibroblasts and muscle cells. It is positive in smooth muscle tumors, glomus tumors, and some atypical fibroxanthomas but is negative in HFLT. This staining pattern is seen with pleomorphic hyalinizing angiectatic tumor (PHAT), which is thought to be a later stage of HFLT.[1,4]

C. $CD34^+$, $desmin^-$ SMA^-, $keratin^-$, $S100^-$ — Correct. HFLT is positive for CD34, an endothelial marker that is also positive in vascular tumors, DFSP, epithelioid sarcoma, spindle cell lipoma, neurofibroma, and others. HFLT is negative for desmin, SMA, keratin, and S100 protein.[3,5]

D. $CD34^-$, $desmin^-$, SMA^-, $keratin^+$, $S100^+$ — Incorrect. Keratin is found in epithelial cells. It is positive in epithelial tumors and some adnexal tumors as well as epithelioid sarcoma, which is

$CD34^+$ and $keratin^+$ (AE1/AE3), but keratin is negative in HFLT.[3]

E. $CD34^-$, $desmin^+$, SMA^-, $keratin^+$, $S100^+$ — Incorrect. Desmin is found in muscle cells (skeletal and smooth muscle) and positive in leiomyomas and leiomyosarcomas but is negative in HFLT.[3]

Question 3: This type of tumor shares an identical chromosomal translocation with which of the following?

A. Malignant fibrous histiocytoma (MFH)

B. Myxoinflammatory fibroblastic sarcoma (MIFS)

C. Leiomyosarcoma

D. Myxoid liposarcoma

E. Atypical fibroxanthoma

Answers:

A. MFH — Incorrect. MFH is a soft-tissue sarcoma that frequently occurs in deep soft tissues of the extremities with a high risk of metastasis, but MFH does not share an identical chromosomal translocation with HFLT.[1]

B. MIFS — Correct. MIFS and HFLT share an identical chromosomal translocation t(1;10)(p22;q24) in a subset of tumors with the fusion gene *TGFBR3-MGEA5*. Further, some HFLT tumors can progress to a so-called *hybrid hemosiderotic fibrolipomatous tumor- myxoinflammatory fibroblastic sarcoma* that is similar to, but distinct from, classic MIFS. PHAT also shares the same identical chromosomal translocation in a subset of patients.[6]

C. Leiomyosarcoma — Incorrect. Leiomyosarcoma is a superficial malignant smooth muscle tumor that does not share an identical chromosomal translocation with HFLT.[1]

D. Myxoid liposarcoma — Incorrect. Myxoid liposarcoma is a type of liposarcoma that often presents as an asymptomatic nodule. This tumor is associated with a t(12;16)(q13;p11) translocation resulting in a FUS/DDIT3 fusion protein (>90% of cases).[1]

E. Atypical fibroxanthoma — Incorrect. Atypical fibroxanthoma is a superficial variant of MFH that typically occurs in elderly patients. Atypical fibroxanthoma does not share an identical chromosomal translocation with HFLT.[1]

Abbreviations:

DFSP: dermatofibrosarcoma protuberans
HFLT: hemosiderotic fibrolipomatous tumor
MFH: malignant fibrous histiocytoma
MIFS: Myxoinflammatory fibroblastic sarcoma
PHAT: pleomorphic hyalinizing angiectatic tumor
SMA: smooth muscle actin

REFERENCES

1. Lindberg MR. *Diagnostic Pathology: Soft Tissue Tumors*. Elsevier Health Sciences; 2015
2. Moretti VM, Brooks JSJ, Ogilvie CM. Case report: hemosiderotic fibrohistiocytic lipomatous lesion: a clinicopathologic characterization. *Clin Orthop Relat Res*. 2010;468(10):2808-2813.
3. Ferringer T. Immunohistochemistry in dermatopathology. *Arch Pathol Lab Med*. 2015;139:23.
4. Kao Y, Ranucci V, Zhang L, et al. Recurrent BRAF gene rearrangements in myxoinflammatory fibroblastic sarcomas, but not hemosiderotic fibrolipomatous tumors. *Am J Surg Pathol*. 2017;41(11):1456-1465.
5. Wilk M, Zelger BG, Zelger B. Hemosiderotic fibrolipomatous tumor. *Am J Dermatopathol*. 2016;38(9):714-716.
6. Boland JM, Folpe AL. Hemosiderotic Fibrolipomatous tumor, pleomorphic hyalinizing angiectatic tumor, and myxoinflammatory fibroblastic sarcoma: related or not? *Adv Anat Pathol*. 2017;24(5):268-277.